

Bladder Exstrophy Girls

What is it?

Bladder Exstrophy is a congenital defect (present at birth) where the bladder is turned "inside out". The bladder does not form into its normal round shape but instead is flattened and exposed outside the body. The lower portion of the bladder, a funnel shaped bladder neck made up of muscles that open and close the bladder, fails to form correctly. The cause of Bladder Exstrophy is unknown.

- ▶ The problem occurs somewhere between 4 – 10 weeks of pregnancy when various organs, tissues and muscles are developing.
- ▶ Bladder Exstrophy is **not** inherited and did **not** occur because of anything the mother did or did not do during pregnancy.

Incidence

- ▶ Bladder Exstrophy is noted in approximately 1 in 18,500 live births.¹
- ▶ Bladder Exstrophy is more likely to occur in boys than girls by approximately 5 to 1.
- ▶ The risk of having a 2nd child with Bladder Exstrophy is about 1 in 100, and 1 in 70 if one of the parents has Bladder Exstrophy.



Reference:

1. EUROCAT (the European Surveillance System for Congenital Anomalies) <http://www.eurocat-network.eu/> @ 31.08.2010. Period covered 2000 - 2008 inclusive.

Signs and Symptoms in Girls

- ▶ Usually the uterus, fallopian tubes and ovaries are normal.
- ▶ The vagina may be narrowed and misplaced.
- ▶ The clitoris is separated into two parts and the urethral opening is between the clitoris.
- ▶ The pelvic bones will be widely separated at the front (pubis).

Investigations may include:

- ▶ ultrasound of kidneys at regular intervals
- ▶ I.V.P.
- ▶ DMSA Scan
- ▶ urodynamics
- ▶ CT scans of the pelvis

Frequent hospital visits will be necessary for your child to be assessed.

Treatment / Surgery

You need to be aware that each child is different, and your doctor may decide which surgery and treatment plan is best for your child. In most cases more than one operation will be necessary and your doctor will discuss this with you.

The following information is generalised and not specific to your child:

- ▶ Closure of bladder and tummy, and bringing together the pubic bones may be undertaken in the first four weeks of life. In specific cases, bladder closure may be performed in the first 24 hours of life, usually in hospital for between 4 – 6 weeks.
- ▶ Reconstructive surgery to the clitoris (1-1½ years). Usually resulting in a hospital stay for one week or longer.



Long Term Management

Some of the following may be necessary:

- ▶ The bladder size may be very small therefore an operation may be needed to make this bigger (augmentation), so more wee can be stored.
- ▶ Most children will need to use a small tube (a catheter) to empty the bladder several times a day – this is called intermittent catheterisation and will be taught by a specialist nurse.
- ▶ Some children may need to have a small opening made on the tummy from the bladder (Mitrofanoff) to be able to pass the catheter to perform intermittent catheterisation.
- ▶ Some children may not wish to use a catheter to empty the bladder. They will be able to have an opening (stoma) made on the tummy to be able to collect the wee in a special bag.
- ▶ Some children may have problems with wee flowing back up the ureters to the kidneys (reflux) – this can cause serious damage to the kidneys and may have to be treated with an operation called ‘re-implantation of ureters’.

Possible Problems

- ▶ Urinary Tract Infections.
- ▶ Latex allergy – occasionally children may develop an allergy to latex which is used to make surgical gloves. As with any allergy this can range from a minor reaction to something more serious. If you are concerned, contact your doctor.
- ▶ Urinary incontinence requiring further surgery to gain continence.



Child's health and development

- ▶ Your child will require frequent visits to hospital and may still be incontinent when starting school. This can affect education but in the majority of cases the children are healthy with normal intelligence, and normal physical and social development.
- ▶ Your child may have a waddling walk which will become less obvious as she gets older.
- ▶ Extra care will be needed for urinary control.
- ▶ When reaching puberty, girls may require genital surgery but should be able to have normal sexual intercourse. Almost all females are able to have children. When ready to start a family, her doctor should be consulted as she may be prone to prolapse of the uterus. This is not dangerous when monitored carefully and the doctor will advise if a caesarean section is thought preferable.

What will happen to us emotionally?

- ▶ You will have to deal with questions from family and friends.
- ▶ When your child reaches school age you will have to deal with questions from other people.
- ▶ Social issues with your child, other children and adults may be difficult.
- ▶ Your life will be different and probably more difficult than those around you.

Support

Support and counselling will be given throughout by your Clinical Nurse

Specialist and he/she will give you relevant information for:

- ▶ support groups – parent led groups
- ▶ financial support
- ▶ educational issues

Please don't hesitate to ask for advice on anything which is worrying you or your family!